Policy Title: Haegarda (C1 Esterase Inhibitor Subcutaneous [Human]) (subcutaneous)

| Effective Date: | 01/01/2020 |
| Review Date: | 09/25/2019, 12/18/19, 1/22/20, 9/9/2020, 5/6/2021 |
| Revision Date: | 09/25/2019, 1/22/20, 9/9/2020, 5/6/2021 |

**Purpose:** To support safe, effective and appropriate use of Haegarda (C1 Esterase Inhibitor Subcutaneous [Human]).

**Scope:** Medicaid, Commercial, Medicare-Medicaid Plan (MMP)

**Policy Statement:**

Haegarda (C1 Esterase Inhibitor Subcutaneous [Human]) is covered under the Medical Benefit when used within the following guidelines. Use outside of these guidelines may result in non-payment unless approved under an exception process.

**Procedure:**

Coverage of Haegarda (C1 Esterase Inhibitor Subcutaneous [Human]) will be reviewed prospectively via the prior authorization process based on criteria below.

**Initial Criteria**

- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements; OR
- Medication is prescribed by, or in consultation with allergist/immunologist or a physician who specializes in the treatment of HAE or related disorders; AND
- Member will not use Haegarda concomitantly with Cinryze, Orladeyo, or Takhzyro.
- Dose does not exceed FDA approved labeling; AND
- Patient has documented diagnosis of HAE type I or type II and meets one of the following:
  - Member has C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing and meets both of the following criteria:
    - Member has a C4 level below the lower limit of normal as defined by the laboratory performing the test, AND
    - Member meets one of the following criteria:
      - C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test, OR
- Normal C1-INH antigenic level and a low C1-INH functional level (functional C1-INH less than 50% or C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test); OR
  - Member has normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria:
    - Member has an F12, angiopoietin-1, plasminogen, or kininogen-1 (KNG1) gene mutation as confirmed by genetic testing, OR
    - Member has a documented family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine (e.g., cetirizine) for at least one month.

*Continuation of Therapy Criteria:*

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria and has documentation of positive clinical response (i.e., decrease in HAE acute attack frequency, decrease in HAE attack severity, or decrease in duration of HAE attacks) since initiating Haegarda prophylactic therapy compared with baseline (i.e., prior to initiating prophylactic therapy).

**Coverage durations:**

- Initial coverage: 6 months
- Continuation of therapy coverage: 6 months

*** Requests will also be reviewed to National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) if applicable. ***

**Dosage/Administration:**

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
<th>Maximum dose (1 billable unit = 10 IU )</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prophylaxis of Hereditary Angioedema (HAE) attacks</td>
<td>60 IU/kg body weight injected subcutaneously twice weekly (every 3 or 4 days)</td>
<td>5,600 billable units per 28 days</td>
</tr>
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**Investigational use:** All therapies are considered investigational when used at a dose or for a condition other than those that are recognized as medically accepted indications as defined in any one of the following standard reference compendia: American Hospital Formulary Service Drug information (AHFS-DI), Thomson Micromedex DrugDex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs, or Peer-reviewed published medical literature indicating that sufficient evidence exists to support use. Neighborhood does not provide coverage for drugs when used for investigational purposes.

**Applicable Codes:**
Below is a list of billing codes applicable for covered treatment options. The below tables are provided for reference purposes and may not be all-inclusive. Requests received with codes from tables below do not guarantee coverage. Requests must meet all criteria provided in the procedure section.

The following HCPCS/CPT code is:

<table>
<thead>
<tr>
<th>HCPCS/CPT Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>J0599</td>
<td>Injection, c-1 esterase</td>
</tr>
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References: